What is claimed is:

1. A transgenic mouse harboring a homozygous null mutation in its endogenous Alpha Hemoglobin Stabilizing Protein (AHSP) gene wherein said mouse does not express a functional mouse Alpha Hemoglobin Stabilizing Protein (AHSP) protein and erythrocytes obtained from said mouse exhibit one or more characteristics selected from the group consisting of abnormal spiculated morphology, reduced lifespan, increased production of reactive oxygen species (ROS), and precipitated hemoglobin.

2. The transgenic mouse of claim 1, wherein said mouse is fertile and transmits said null mutation to its offspring.

3. The transgenic mouse of claim 1, wherein said null mutation has been introduced into an ancestor of said mouse at an embryonic stage following microinjection of embryonic stem cells into a mouse blastocyst.

4. The transgenic mouse of claim 1, wherein said null mutation has been introduced into an ancestor of said mouse at an embryonic stage following co-incubation of embryonic stem cells with a fertilized egg or morula.

5. A transgenic mouse harboring a heterozygous mutation in its endogenous Alpha Hemoglobin Stabilizing Protein (AHSP) gene wherein said mouse exhibits AHSP haploinsufficiency and has an elevated reticulocyte count.

6. The transgenic mouse of claim 5, wherein said mouse is fertile and transmits said mutation to its offspring.

7. The transgenic mouse of claim 5, wherein said mutation has been introduced into an ancestor of said mouse at an embryonic stage following microinjection of embryonic stem cells into a mouse blastocyst.

8. The transgenic mouse of claim 5, wherein said mutation has been introduced into an ancestor of said mouse at an embryonic stage following co-incubation of embryonic stem cells with a fertilized egg or morula.

9. A method for screening for therapeutic agents which affect Alpha Hemoglobin Stabilizing Protein (AHSP) activity, comprising:

a) administering a test compound to the transgenic mouse of claim 1;

b) assessing said mouse for an alteration in an Alpha Hemoglobin Stabilizing Protein (AHSP) activity.

10. The method of claim 9, wherein said activity is selected from the group consisting of α-hemoglobin binding, and α-hemoglobin synthesis.

11. A method for assessing the activity of a compound useful for the treatment and/or prevention of an AHSP related disorder, comprising:

a) providing at least one Alpha Hemoglobin Stabilizing Protein (AHSP) knock-out mouse;

b) administering a test compound to said mouse; and

C) assessing said mouse for inhibition of said AHSP related disorder.

12. The method of claim 11, said method further comprising administration of said test compound to a control mouse to assess toxicity of said test compound.

13. The method of claim 11, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

14. A method of diagnosing an Alpha-Hemoglobin Stabilizing Protein (AHSP) related disorder in a test subject, wherein a sample from said test subject is analyzed by a method selected from the group consisting of:

a) a method employing a specific binding member capable of binding to a AHSP nucleic acid sequence, the specific binding member comprising nucleic acid hybridizable with the AHSP sequence; and

b) a method of determining the presence, in a sample from a test subject, of a polypeptide encoded by the AHSP nucleic acid and, if present, determining the expression level; and

c) a method wherein at least one antibody domain with specificity for an epitope selected from the group consisting of a native AHSP nucleic acid sequence epitope, or a polypeptide epitope, the specific binding member being labeled so that binding of the specific binding member to its binding partner is detectable; and

d) a method of PCR amplification involving one or more primers based on AHSP gene sequence to screen for an decrease in AHSP expression in a sample from a test subject; and

e) a method of determining the presence, in a sample from a test subject, of a polypeptide encoded by the AHSP

nucleic acid and, if present, determining the presence of mutations of the AHSP nucleic acid.

15. The method of claim 14, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

16. The method of claim 15, wherein said disorder is bovine spongiform encephalopathy.

17. A method of screening for compounds which modulate the activity of an AHSP polypeptide, the method comprising contacting at least one test compound with the AHSP polypeptide in a reaction medium, testing the activity of the treated AHSP polypeptide and comparing that activity with the activity of native, untreated AHSP polypeptide in a comparable reaction medium.

18. A compound identified by the method of claim 11 or 17, wherein said compound is a fragment of AHSP, or a small molecule which mimics AHSP activity.

19. A method of treating, or ameliorating symptoms of an AHSP related disorder comprising over expressing an AHSP encoding nucleic acid molecule in the cells or body fluid of a patient having said disorder.

20. The method of claim 19, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

21. A method of treating, or ameliorating symptoms of an AHSP related disorder comprising administering the compound of claim 18 to the cells or body fluid of a patient having said disorder.

22. The method of claim 21, wherein said disorder is selected from the group consisting of α-thalassemia, β-thalassemia, anemia, spongiform encephalopathy, prion disease, and Alzheimer's disease.

23. A method for producing alpha hemoglobin stabilizing protein (AHSP)-specific antibodies, comprising:

a) immunizing an AHSP knock out mouse with an immunogenic amount of AHSP or fragments thereof;

b) harvesting serum from said mouse; and

c) screening said serum for antibodies immunoreactive to AHSP.

24. An antibody preparation produced by the method of claim 23.

25. A method for producing alpha hemoglobin stabilizing protein (AHSP)-specific antibodies, comprising:

a) immunizing an AHSP knock out mouse with an immunogenic amount of AHSP or fragments thereof;

b) harvesting the spleen of said mouse and fusing said spleen cells with a myeloma cell line containing a mutation to facilitate isolation of fused spleen/myeloma cells;

c) culturing said fused cells in media containing a selection agent wherein fused cells grow, and non-fused cells are killed;

d) screening said media from cells surviving in the presence of said selection agent, for the presence of antibodies immunoreactive to AHSP; and

e) optionally isolating said antibody.

26. A monoclonal antibody produced by the method of claim 25.

27. A kit comprising one or more molecules for detecting AHSP expression, said molecules being optionally detectably labeled, and said molecules being selected from the group consisting of nucleic acid molecules having sequences corresponding to a portion of an AHSP nucleic acid sequence for use in amplifying a nucleic acid comprising an AHSP nucleic acid sequence, and antibodies which specifically bind to a portion of the AHSP protein.

28. A transgenic mouse characterized by overexpression of an Alpha Hemoglobin Stabilizing Protein (AHSP) gene.

29. The transgenic mouse of claim 1, said mouse also having a heterozygous null mutation in beta major and minor globulin genes on a single chromosome, said mouse exhibiting at least one characteristic selected from the group consisting of low hematocrit level, and increased red cell distribution width (RDW).

30. A method for assessing the activity of compounds useful for the treatment and/or prevention of an AHSP related disorder, comprising:

a) providing mice as claimed in claim 29;

b) administering a test compound to the mice of step (a); and

C) assessing said mice for inhibition of said AHSP related disorder.

31. The method of claim 30, said method further comprising administration of said compound to control mice to assess toxicity of said test compound.

32. A compound identified by the method of claim 30.

33. A method of treating, or ameliorating symptoms of an AHSP related disorder comprising administering the compound of claim 32 to the cells or body fluid of a patient having said disorder.

34. The transgenic mouse of claim 1, said mouse also having a homozygous null mutation in at least one alpha globulin gene, said mouse exhibiting at least one characteristic selected from the group consisting of low hematocrit level, high reticulocyte count, decreased mean corpuscular volume (MCV), and increased red cell distribution width (RDW).

35. A method for assessing the activity of compounds useful for the treatment and/or prevention of an AHSP related disorder, comprising:

a) providing mice as claimed in claim 34;

b) administering a test compound to the mice of step (a); and

C) assessing said mice for inhibition of said AHSP related disorder.

36. The method of claim 35, said method further comprising administration of said compound to control mice to assess toxicity of said test compound.

37. A compound identified by the method of claim 35.

38. A method of treating, or ameliorating symptoms of an AHSP related disorder comprising administering the compound of claim 37 to the cells or body fluid of a patient having said disorder.